Pediatr Radiol (1997) 27: 339–341

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An extensive type of polyostotic fibrous dysplasia

Accepted: 12 January 1996

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Abstract The case describes an 11-year-old girl affected by an unusually extensive type of polyostotic fibrous dysplasia. The cranium and face and both femurs, tibias, and fibulas were extensively and almost symmetrically involved. Tubulation deformities were noted in the metacarpals and middle phalanges of both hands. These findings appear to represent a very severe manifestation of the disease, as polyostotic fibrous dysplasia is known to be predominantly unilateral. The patient had endocrine dysfunction consistent with McCune-Albright syndrome. Radiological work-up included plain radiography of the skeleton and CT, MR imaging, and MR angiography of the cranium.

Introduction

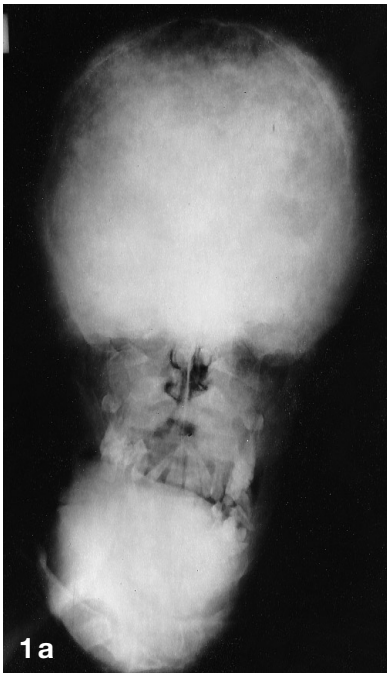
Fibrous dysplasia presents in either a monostotic or a polyostotic form. The polyostotic form of the disease is characterized by predominantly unilateral involvement, and in association with an endocrine dysfunction it is referred to as McCune-Albright syndrome [1–3].

Case report

The present patient is an 11-year-old girl affected by an unusual type of polyostotic fibrous dysplasia. The cranium and face and both femurs, tibias, and fibulas were extensively and almost symmetrically involved (Fig. 1). The pelvis and spine were less involved. The patient's upper extremities were almost spared; however, tubulation deformity of her hand bones was evident. She had multiple, irregular hyperpigmentations of the skin and sexual precocity, identifying the condition as McCune-Albright syndrome. She had undergone two partial surgical resections for a large mandibular mass. Histopathological examination of the specimens revealed a well-vascularized, cellular fibrous tissue consisting of spindle cells, osteocytes and osteoblasts associated with immature woven bone and an abnormal trabeculation pattern, establishing the diagnosis of fibrous dysplasia. No evidence of malignancy was found. On CT a thickened and coarsened calvaria was seen. None of the cranial and facial bones was spared, and all the air sinuses were obliterated. A cranial MR examination revealed a mixed signal pattern of the thickened bones. The abnormal tissue had a generalized hypointense signal on T1- and T2-weighted images, which was more prominent in the latter. The abnormal tissue included scattered nodular or coalescent foci of hyperintensity on T1-weighted images (Fig. 2a, b). Some of these became brighter on T2-weighted images, while others got darker (Fig. 2c, d). The brain was grossly normal (Fig. 2), except for apparent bifrontal impressions from the expanded (as thick as 5 cm) frontal bone (Fig. 2b, c). Surprisingly, no clinically detectable cranial nerve involvement was noted, and the neural exit foramina appeared grossly normal on CT. An MR angiographic study was also undertaken which revealed normal intracranial vascular structures and prominence of extracranial vessels (external carotid artery branches and superficial veins). A number of vessels were identified traversing the thickened calvaria (Fig. 3).

Discussion

Fibrous dysplasia is a developmental osseous disorder of unknown etiology in which cancellous bone is replaced with fibrous tissue, resulting in expansion of the bone. The disease presents in either a monostotic or a polyostotic form. The polyostotic form of the condition in association with skin pigmentation and endocrine abnormalities leading to accelerated maturation and sexual precocity is known as McCune-Albright syndrome. Cranial involvement is seen in most of those affected, and fibrocystic changes in the mandible are particularly noted. With respect to extracranial involve-



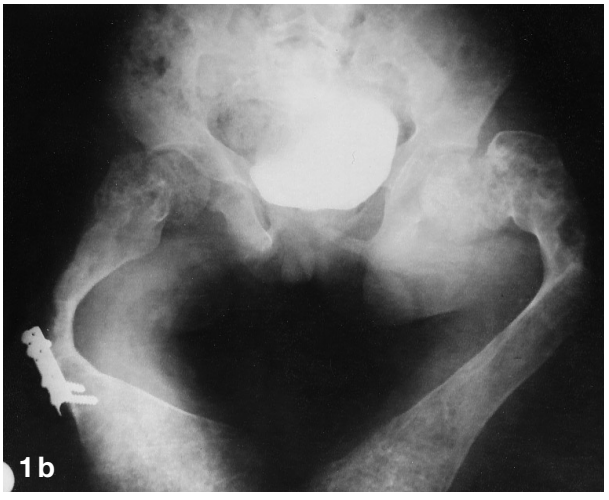
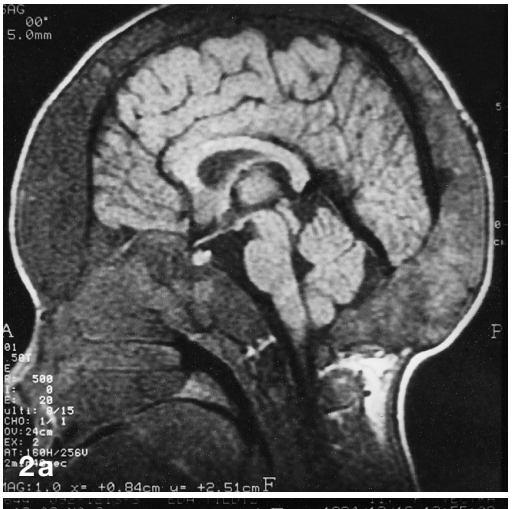
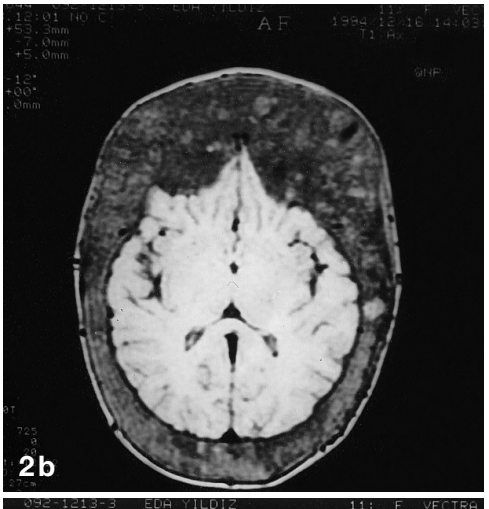
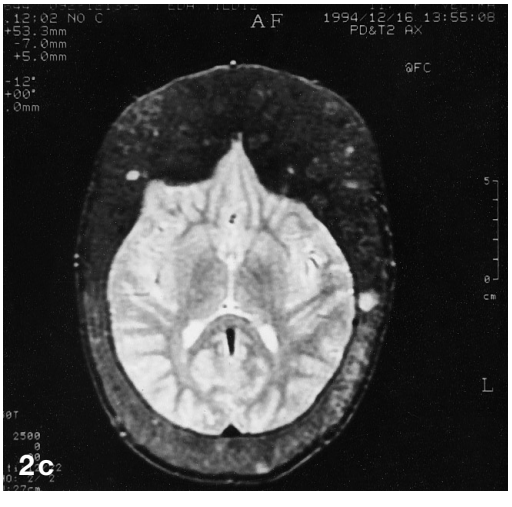


Fig. 1 a, b Radiographs of **a** the cranium and **b** the pelvis and femurs show extensive fibrous dysplasia. Note that the lesions are almost symmetrical, an unusual manifestation of polyostotic fibrous dysplasia







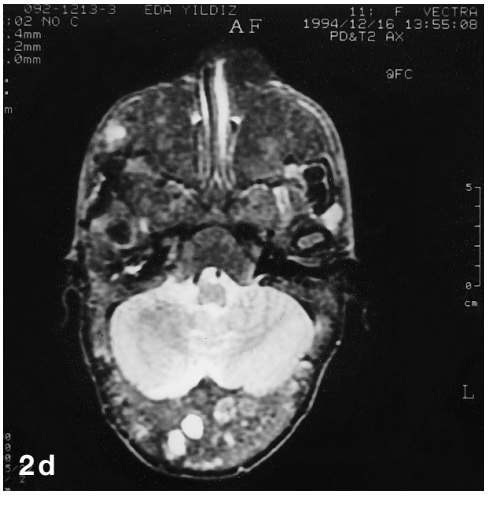


Fig. 2 a, b T1-weighted MR images in the sagittal plane (**a**) and the axial plane (**b**); **c, d** T2-weighted MR images in the axial plane. The lesion has a generalized hypointense signal on T1- and T2-weighted images, more prominent in the latter (**c**). Note scattered nodular or coalescent foci of hyperintensity on T1-weighted images (**a, b**). Some of these become brighter on T2-weighted images, while others get darker (**c**). Note nodules with very high signal in the occipital bone (**d**). The brain is grossly normal except for apparent bifrontal impressions from the expanded (as thick as 5 cm) frontal bone (**b, c**)

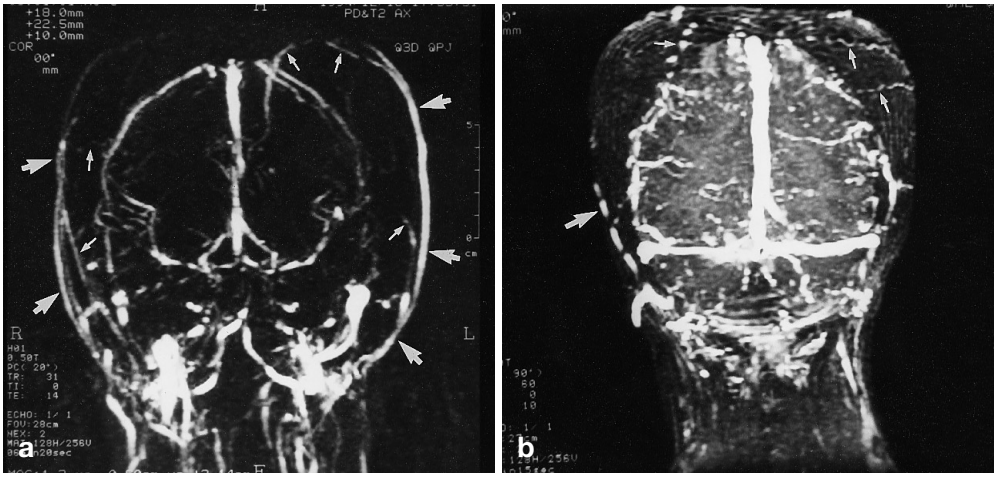


Fig. 3 a, b Coronal maximum intensity projections from MR angiograms obtained at 0.5 T utilizing **a** the three-dimensional phase-contrast technique (TR 31/TE 14, flip angle 20°) and **b** the two-dimensional time-of-flight technique (TR 60/TE 10, flip angle 90°). Note prominent extracranial vessels (external carotid artery branches and superficial veins; *arrowheads*). A number of vessels traverse the thickened calvaria *(arrows)*. Intracranial vessels appear normal

ment, the polyostotic variant usually manifests with unilateral predominancy, and bone deformities are common [1–8]. In the light of these data, it appears that our patient manifests an unusual, extensive type of polyostotic fibrous dysplasia featuring an almost symmetrical involvement of the skeleton and generalized involvement of the face and skull.

It has been reported that fibrous dysplasia demonstrates a predominantly decreased signal (similar to skeletal muscle) within the purely fibrous tissue on both T1- and T2-weighted images. Because the tissue affected is not homogeneous histologically, an inhomogeneous, high signal pattern can be seen within the fibrous tissue on both T1- and T2-weighted MR images, which probably reflects protein-rich parts of the lesion and cartilaginous nodules. On T2-weighted images areas of increased signal are often noted, which may correspond to protein-rich areas or, occasionally, true cysts (very high signal) [4]. The lesions may show enhancement on MR after administration of contrast medium. The reason for this is unknown; the enhancement pattern does not correlate well with the activity of clinically and pathologically determined lesions [4, 5].

In our patient, cranial MR imaging findings were consistent with these reports. We also noted scattered nodular or coalescent foci of hyperintensity on T1-weighted images, some of which became brighter on T2-weighted images, while others got darker, probably reflecting an inhomogeneous fibrous tissue consisting of abnormal trabeculae and cystic changes with high protein content. It is known that fibrous dysplasia appears as a well-vascularized abnormal tissue with numerous small vessels in the center of the lesion and large peripheral sinusoids [4–8]. Our MR angiography findings included prominent extracranial vessels and a number of vessels which traversed the thickened calvaria. Intracranial vascular structures were normal.

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